CASE REPORT

Multiple supratentorial subependymomas causing obstructive hydrocephalus

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SUMMARY

Introduction Subependymomas are benign intraventricular tumours that most often occur asymptotically and are found incidentally on autopsy. Symptomatic examples requiring surgical intervention are exceedingly rare.**Case presentation** A 55-year-old man with no history of neurological symptoms presented with multiple episodes of loss of consciousness and increasing headaches. MRI revealed a lobulated intraventricular mass centred at the right Foramen of Monro. Obstructive hydrocephalus with localised midline shift and a second lesion were noted. Right frontal craniotomy with total removal via transcortical resection was performed.**Discussion** Symptomatic subependymomas generally present with signs of hydrocephalus due to obstruction of cerebrospinal fluid pathways. There is only one other reported case of multifocal subependymomas in a symptomatic patient. An example of multiple supratentorial subependymomas causing obstructive hydrocephalus has not been previously reported.**Conclusions** Multiple subependymomas are rare. Judicious surgical management with full excision led to symptomatic improvement in our patient.

BACKGROUND

Obstructive hydrocephalus can be a devastating neurological disorder that if not recognised and treated appropriately can lead to a significant decrease in a patient's quality of life or even death. Symptomatic subependymomas causing hydrocephalus are rare but do occur and proper management and treatment are required to ensure a positive outcome. Here, we present a case of multiple subependymomas causing obstructive hydrocephalus, which has not been previously reported. We also show that judicious surgical management including full excision of both lesions can lead to complete neurological recovery.

CASE PRESENTATION

A 55-year-old man with no history of neurological symptoms presented with multiple episodes of loss of consciousness and increasing headaches over a 1-year period of time.

INVESTIGATIONS

MRI revealed a lobulated intraventricular mass (measuring 4.1×3.0×3.2 cm) centred at the right Foramen of Monro, with minimal contrast enhancement (figure 1A–C—axial, sagittal and

coronal, respectively). Obstructive hydrocephalus with localised midline shift was noted. A second smaller (9 mm) lesion was also observed along the midbody of the right lateral ventricle (seen posterior to the larger lesion and along the lateral wall of the right lateral ventricle on figure 1A).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of a minimally enhancing intraventricular tumour seen on MRI includes subependymoma, subependymal giant cell astrocytoma, subependymal tubers, astrocytoma and central neurocytoma. The lesions listed in this differential diagnosis can often begin to be differentiated based on clinical factors such as age and location within the ventricular system; however, imaging characteristics are often non-specific: subependymoma (these are more often identified in adults and even incidentally at autopsy, location is often in the frontal horn or body of the lateral ventricle but they can also be found in the fourth ventricle), subependymal giant cell astrocytoma (occurs in older children and young adults with tuberous sclerosis, location is typically at the Foramen of Monro), subependymal nodules in tuberous sclerosis (these are found in children and young adults with tuberous sclerosis, location for these lesions is typically near the caudate nucleus along the striothalamic groove or less commonly in the atria or temporal horns of the lateral ventricles), astrocytoma (can be found in children and adults, location is typically along the septum pellucidum, near the Foramen of Monro within the lateral or third ventricle) and central neurocytoma (usually diagnosed between 20 and 40 years of age, location is within the body of the lateral ventricle).¹

Although treatment for each of these conditions is surgical, proper histopathological diagnosis is critical for patient counselling and outcome expectation. Differential diagnosis from a histopathological viewpoint includes subependymoma (typical histological bland nuclei in ill-defined clusters amidst small microcysts), subependymal nodules or subependymal giant cell astrocytoma (unique to patients diagnosed with tuberous sclerosis), astrocytoma and central neurocytoma (typical histological salt and pepper appearance but lack 1p19q as seen in oligodendrogliomas).

In our case, proper identification of subependymomas allowed for accurate counselling regarding the benign nature and non-progressive course our patient could expect.

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Figure 1 (A) Contrast-enhanced T1-weighted axial MRI demonstrating two intraventricular lesions, the larger lesion extends from the septum pellucidum and measures 4.1×3.0×3.2 cm and causes obstruction of cerebrospinal fluid flow at the Foramen of Monro. The smaller lesion extends from the midbody of the right lateral ventricle and measures 9 mm. (B) Preresection contrast-enhanced T1-weighted sagittal MRI showing the larger intraventricular tumour at the Foramen of Monro. (C) Preresection contrast-enhanced T1-weighted coronal MRI showing the larger intraventricular tumour at the Foramen of Monro with dilation of the frontal horns of the lateral ventricles bilaterally. (D) Postresection contrast-enhanced T1-weighted axial MRI corresponding to the same level as that showing the two tumours prior to resection—also note the decompressed lateral ventricles after re-establishment of normal cerebrospinal fluid flow at the Foramen of Monro. (E) Postresection contrast-enhanced T1-weighted sagittal MRI showing resection of the tumour from the area of the Foramen of Monro (corresponds to the same location depicted in B). (F) Postresection contrast-enhanced T1-weighted coronal MRI showing resection of the tumour from the area of the Foramen of Monro and showing decompression of the frontal horns of the lateral ventricles bilaterally (corresponds to the same location depicted in C)—also note the transcortical resection pathway between the superior and middle frontal gyri.

TREATMENT

A right frontal craniotomy for transcortical resection of both intraventricular tumours was performed through the superior frontal gyrus (figure 2A and B—preresection and postresection, respectively). Both tumours were completely resected without complication, and the patient suffered no neurological deficits postoperatively (figure 1D–F—axial postresection, sagittal postresection

and coronal postresection, respectively, all corresponding to the preresection images in figures 1A–C).

Histopathological examination of both tumours revealed fibrillar lesions of low cellularity. The component cells have small, elliptical, cytologically bland nuclei and occur in small, ill-defined clusters scattered amidst small microcysts. Occasional cells showed some nuclear atypia (large and irregular in shape),

Figure 2 Intraoperative microscope photographs demonstrate (A) a large intact subependymoma attached to the septum pellucidum with visible patty on right and (B) postresection.

but there was no discernible mitotic activity and no other histological features to suggest malignancy (figure 3).

OUTCOME AND FOLLOW-UP

The patient remains neurologically intact at 6 months and 1 year follow-up.

DISCUSSION

Subependymomas are tumours first described by Scheinker in 1945.² Many examples are small lesions found incidentally at autopsy, while symptomatic examples requiring surgical resection are exceedingly rare. They are slow growing, benign neoplasms of uncertain origin most often located in the floor of the fourth ventricle,^{3,4} although they can be found in the lateral ventricles as well.^{5–7} Subependymomas most commonly remain asymptomatic and tend to occur in older individuals (around 60 years of age). However, if symptomatic, the most common presenting symptoms are associated with hydrocephalus (eg, headache, nausea, vomiting and/or ataxia) and presentation tends to be earlier (around 40 years of age).^{3–6}

Subependymomas are believed to comprise between 0.2% and 0.7% of intracranial neoplasms worldwide and affect males more often than females.³ They are rare tumours that are most frequently asymptomatic and found incidentally at autopsy or on neuroimaging completed for other medical indications.^{3,4,6,8} Symptomatic patients are most likely to present with signs of hydrocephalus due to the obstruction of the cerebrospinal fluid pathways and as such patients may present with headache, nausea, vomiting and/or ataxia.^{3–6}

Figure 3 Microscopic examination shows fibrillar lesions of low cellularity. Cells have small, elliptical, cytologically bland nuclei and occur in small, ill-defined clusters scattered amidst small microcysts.

Learning points

- ▶ Subependymomas are generally solitary benign intraventricular lesions.
- ▶ They are most commonly asymptomatic at discovery.
- ▶ Subependymomas can present as multiple concurrent lesions and can occur in a variety of locations within the cerebral ventricular system.
- ▶ Their most common symptomatic presentation is from obstructive hydrocephalus.
- ▶ Obstructive hydrocephalus commonly presents with headache, nausea, vomiting and/or ataxia.

The likelihood of symptomatic presentation is generally felt to be largely based on location and size of the lesion. On the basis of previous reports, tumours larger than 4 cm as well as those located on the septum pellucidum or at the Foramen of Monro were the most likely to become symptomatic.^{3,4} However, another more recent retrospective study by Rushing *et al*⁹ evaluated 83 cases of subependymoma and found no correlation between tumour size or location and clinical behaviour.⁹ While not present in this case, one other potential risk factor that may lead to a symptomatic presentation for subependymoma is tumour-associated haemorrhage.¹⁰

Although the vast majority of subependymomas remain asymptomatic and therefore go undetected, those that are symptomatic can be treated by total surgical resection. Total resection is most successfully accomplished for supratentorial subependymomas as they tend to be well demarcated, non-invasive and avascular.^{5,6} Previous studies have found that perioperative mortality rates could be as high as 51% in these patients, although more recent studies suggest a lower mortality rate (23.5%).⁹

The role for postsurgical radiation in those requiring treatment is significantly debated, as surgical resection alone is often curative.^{4,6} However, there are reports of successful adjuvant management by radiation therapy in rare recurrent cases, as well as in cases where subtotal resection was performed.¹¹

A current review of the literature revealed over 100 reported cases of symptomatic supratentorial subependymomas, none of which included multiple lesions.^{4–7,9,12–23} To date, we are aware of only one other reported case of multifocal subependymomas in a symptomatic patient—who presented with three infratentorial subependymomas.²⁴ Therefore, a finding of multiple supratentorial subependymomas presenting symptomatically is unprecedented.

Contributors JS managed the patient clinically and worked extensively in the article preparation and review. DLF and PE were involved in patient care and initial and subsequent manuscript writing. REM performed pathological examination and diagnosis and wrote the histopathological description. All authors have reviewed and approved the manuscript and have met the requirements recommended by the ICMJE for full authorship.

Competing interests None declared.

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